Rheumatic Fever

Ahmed Rashad Mashaal

DEFINITION:

- Acute rheumatic fever is a systemic disease of childhood ,often recurrent that follows group A beta hemolytic streptococcal infection
- It is a diffuse inflammatory disease of connective tissue primarily involving heart, blood vessels, joints, subcut.tissue and CNS

Etiology

- Beta hemolytic streptococci.
- Genetic

Epidemiology

- Ages 5-15 yrs are most susceptible
- Rare <3 yrs
- Girls>boys
- Environmental factors--*over crowding, poor sanitation, poverty,*
- *Incidence more during fall ,winter & early spring*

Pathogenesis

- Delayed immune response to infection with group A beta hemolytic streptococci.
- After a latent period of 1-3 weeks, antibody induced immunological damage occur to *heart valves, joints, subcutaneous tissue & basal ganglia of brain.*

Pathologic Lesions

- Fibrinoid degeneration of connective tissue, inflammatory edema, inflammatory cell infiltration & proliferation of specific cells resulting in formation of **Ashcoff nodules,** resulting in-

  -***Pancarditis*** *in the heart*

  -***Arthritis*** *in the joints*

  -***Ashcoff nodules*** *in the subcutaneous tissue*

  -Basal gangliar lesions resulting in ***chorea***

Pathogenesis

- There is no direct invasion to the tissue by the microorganism but its an auotoimmune disease that involves Ag-Ab interaction.
- It is due to an immunologic reaction that is a delayed sequela of group A betahemolytic streptococcal infections of the pharynx.
- It must be pharyngeal infection not skin infection.

- **Group A streptococcal(GAS) pharyngeal infection**
- Body produce **antibodies** against streptococci ->
- These antibodies **cross react with human tissues** because of the **antigenic similarity** between streptococcal components and human connective tissues (molecular mimicry)[there is certain amino acid sequence that is similar between GAS and human tissue]->
- **Immunologically mediated inflamation & damage (autoimmune)** to human tissues which have antigenic similarity with streptococcal components- like heart, joint, brain connective tissues.

Pathogenesis

Jones Criteria (Revised) for Guidance in the Diagnosis of Rheumatic Fever*

Major Manifestation	Minor Manifestations		Supporting Evidence of Streptococal Infection
	Clinical	Laboratory	
Carditis Polyarthritis Chorea Erythema Marginatum Subcutaneous Nodules	Previous rheumatic fever or rheumatic heart disease Arthralgia Fever	Acute phase reactants: Erythrocyte sedimentation rate, C-reactive protein, leukocytosis Prolonged P-R interval	Increased Titer of Anti-Streptococcal Antibodies ASO (anti-streptolysin O), others Positive Throat Culture for Group A Streptococcus Recent Scarlet Fever

<u>Diagnosis of Rheumatic fever requires:</u>

a) Two major criteria or 1 major and 2 minor criteria

Plus

b) Evidence of recent streptococcal infection (ASOT / throat swab & culture).

Recommendations of the American Heart Association

Major criteria

1- Arthritis

- Inflammation of big joints as ankles, knees and elbows. The joint is:

1. Swollen
2. Hot
3. Tender
4. limited movement
5. Painful

- The inflammation migrates from the affected joint to another leaving the joint normal without damage. The inflammation responds dramatically to salicylates.

Major criteria

2- Carditis

- All layers of the heart are affected (pancarditis)

a- **Endocarditis:**

i. Acutely; murmurs of the blood flow through the narrowed oedematous valves.

ii. Chronic stages: healing by fibrosis, leading to valvular stenosis or regurgitation (left sided valves are the most affected: mitral and aortic)

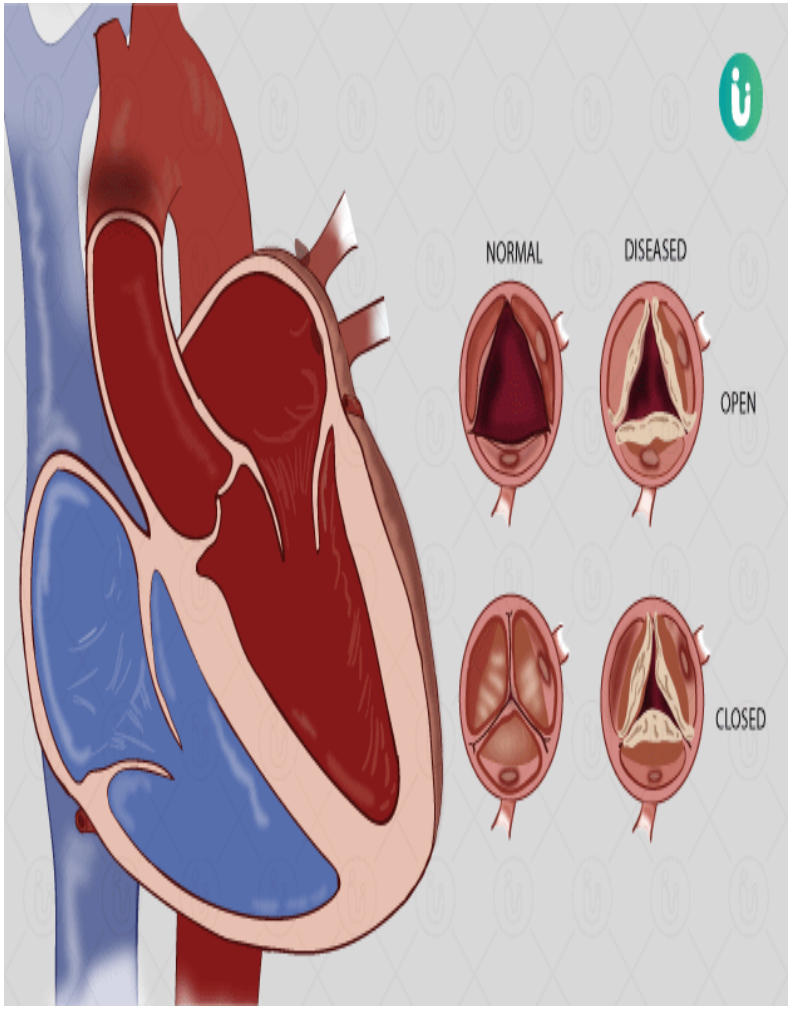
b- **Myocarditis:**

i. Tachycardia

ii. Heart failure (Gallop rhythm).

iii-Cardiac enlargement.

Iv- Arrhythmia.



c- **Pericarditis:**

i-causes stitching chest pain.

ii- May be complicated by pericardial effusion.

Major criteria

3- Chorea

- Termed: Sydenham's Chorea.
- Involuntary, purposeless, unintensional, spasmodic, jerky movement due to affection of the basal ganglia.
- Deterioration of the hand writing, emotional liability, grimacing in the face.
- Pronator sign of the arm, milking sign of the hands.
- Affects mainly young females (1-15years).
- In 10 to 30% of patients.
- Self limited with no long term sequel.

Major criteria

4- Subcutaneous modules

- Painless transient, pea sized, subcutaneous nodules.
- Occur on extensors of large joints and bony prominences.
- Occurs in 10% of patients and associated with strong seropositivity.

5- Erythema marginatum:

- Transient flat painless pink rash mainly on trunk and limbs

Minor criteria

Clinical minor

a. Fever (38.2 – 38.9 c)

b. Arthralgia (joint pain)

c. Pallor

d. Anorexia, weight loss

e. history of previous RF or rheumatic heart disease.

Lab minor

a. Erythrocyte sedimentation rate (ESR) increased

b. Positive C reactive protein

c. Leucocytosis and/or anaemia.

d. ECG shows prolonged P-R interval (first degree heart block).

Management

Prophylactic

1. **Primary prophylaxis**: proper treatment of repeated attacks of pharyngitis and tonsillitis. Tonsillectomy if needed.
2. **Secondary prophylaxis**: to prevent recurrent attacks all patients should receive long acting penicillin, Benzathine penicillin 1.2 million unit deep IM every 3-4 weeks for life, age of 25 or al least for 5 years after the last attack. Allergic patients should receive erythromycin 250 mg/6 hours daily.

Therapeutic

1- Bed rest

2-Phynoxymethyl penicillin 500 mg orally / 6 hours for 8 days

3-Salicylate high dose for arthritis

4-Aspirin and corticosteroids for carditis

Valvular heart diseases and infective endocarditis

- Diseases that affect the mitral, aortic, pulmonary and tricuspid valves may result in stenosis or regurgitation.
- After 10-20 years of acute rheumatic fever with carditis, more than 50% of patients develop rheumatic valve disease affecting mainly the mitral and aortic valves.
- Other causes of valve diseases are: congenital, functional, calcific, collagen vascular disorders and others.
- More than one valve may be affected in the same patient (e.g.) mitral stenosis and aortic regurge.
- The same valve may be affected with two lesions (e.g.) mitral stenosis and regurge (double mitral).

Complications of the valvular heart diseases:

- Heart failure.
- Infective endocarditis.
- Arrhythmias especially atrial fibrillation.
- Arterial embolisation.

Infective Endocarditis

(subacute bacterial endocarditis)

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Introduction

Definition

- A serious bacterial infection of the endocardium (including heart valves).
- Caused by colonization and invasion to the heart valve (or mural endocardium) by microbe.
- The heart valve affected is either native (normal or diseased) or prosthetic.
- Leading to formation of large friable vegetations, which is composed from thrombotic debris (fibrin + Platelets) & the organisms.
- Often lead to destruction of the affected cardiac tissue.

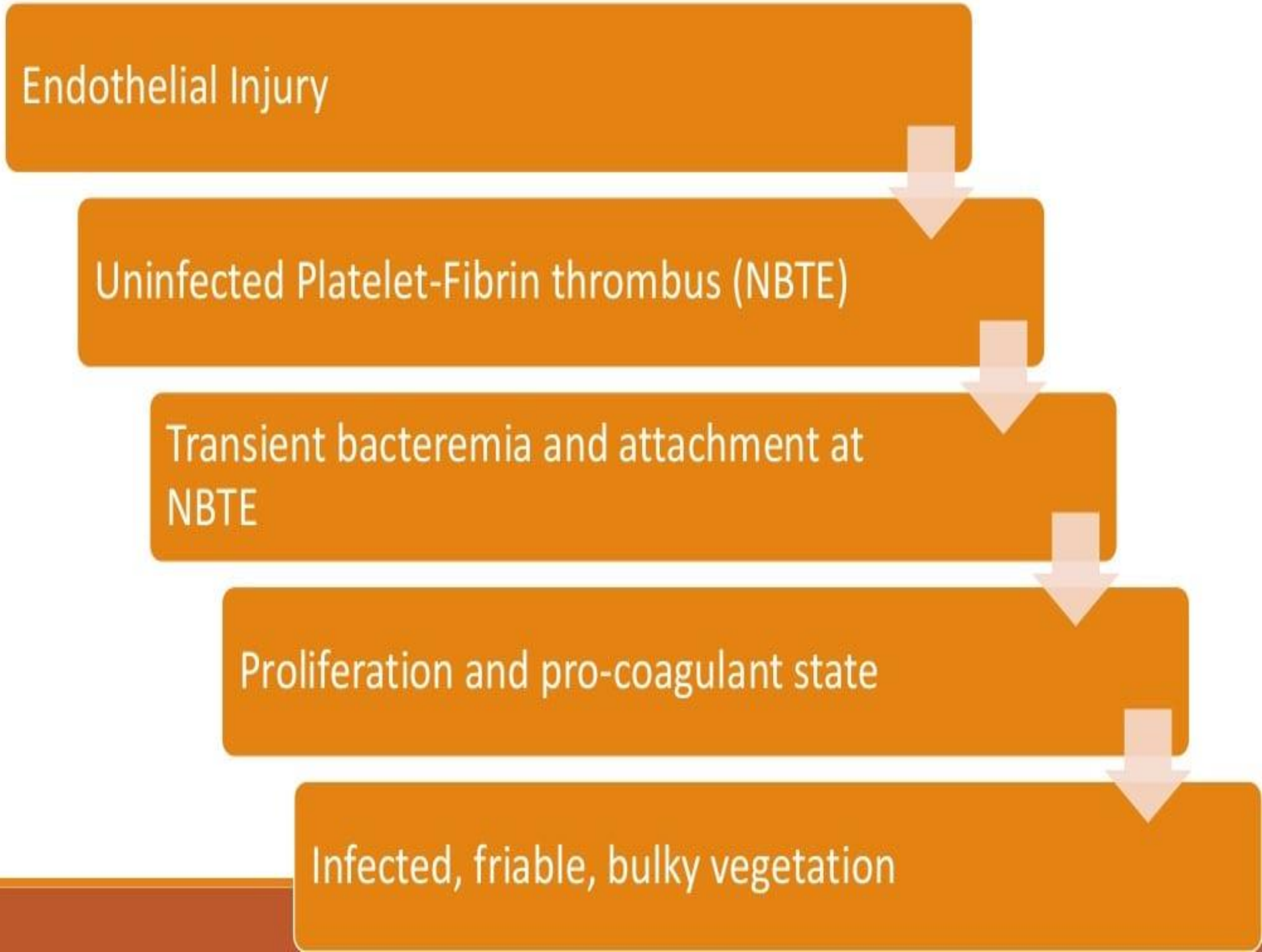
Pathogenesis 1

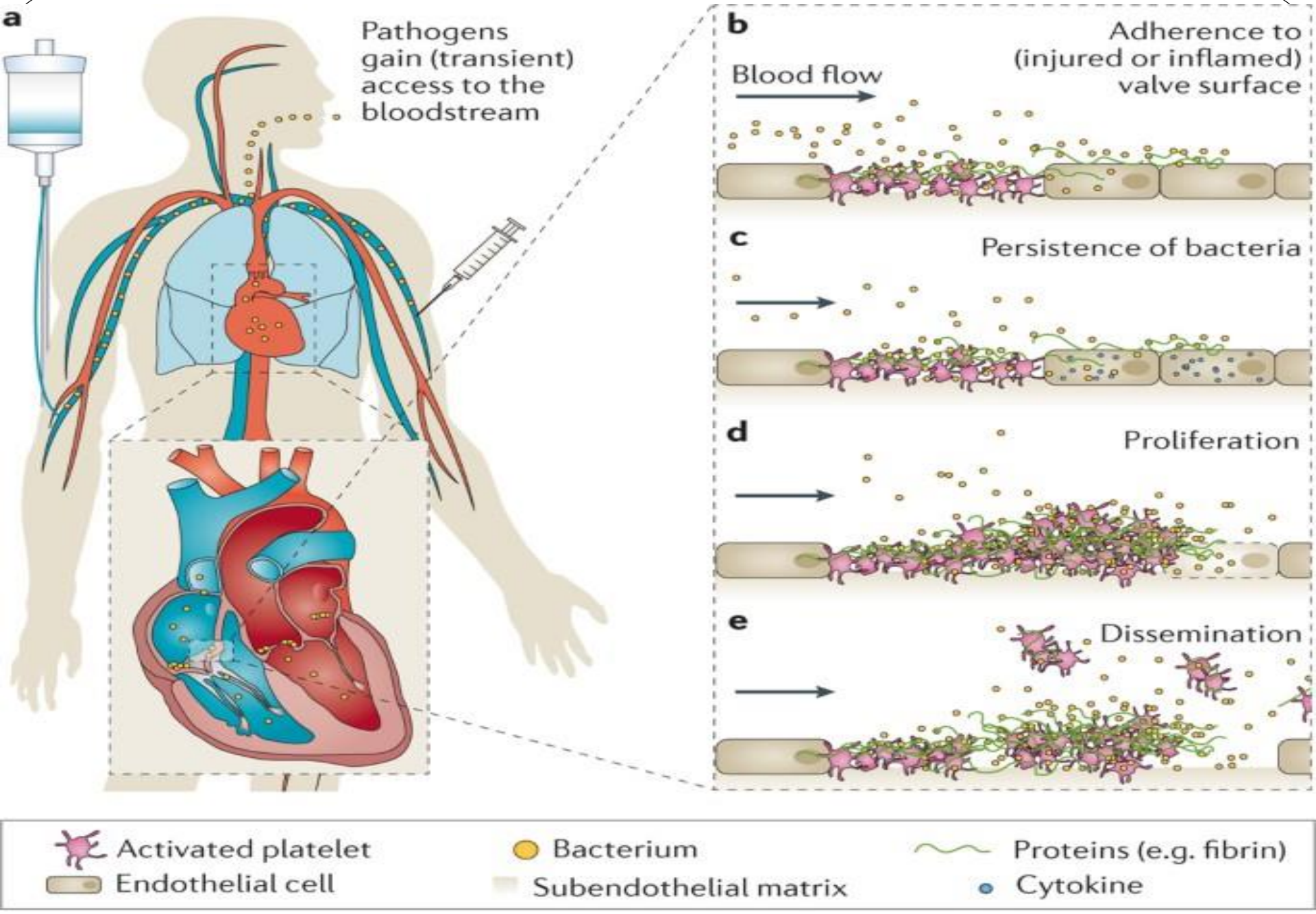
Microbe

1. *Streptococcus viridans (which are normal oral flora) pass to circulation causing transient bacteremia, following dental procedures as: Tooth extraction, Surgery, Scaling, Instrumentation beyond the root apex during endodontic treatment.*
2. *Streptococcus (enterococci) feacalis may give the same effects following bowel and urinary tract instrumentation, surgery or urinary catheterization.*
3. *Staphylococcus aurius may be also the cause if reach circulation in drug abusers and intravenous catheters.*
4. *Others: Streptococcal epidermidis– Gram negative bacilli of HACEK – yeast & fungi.*

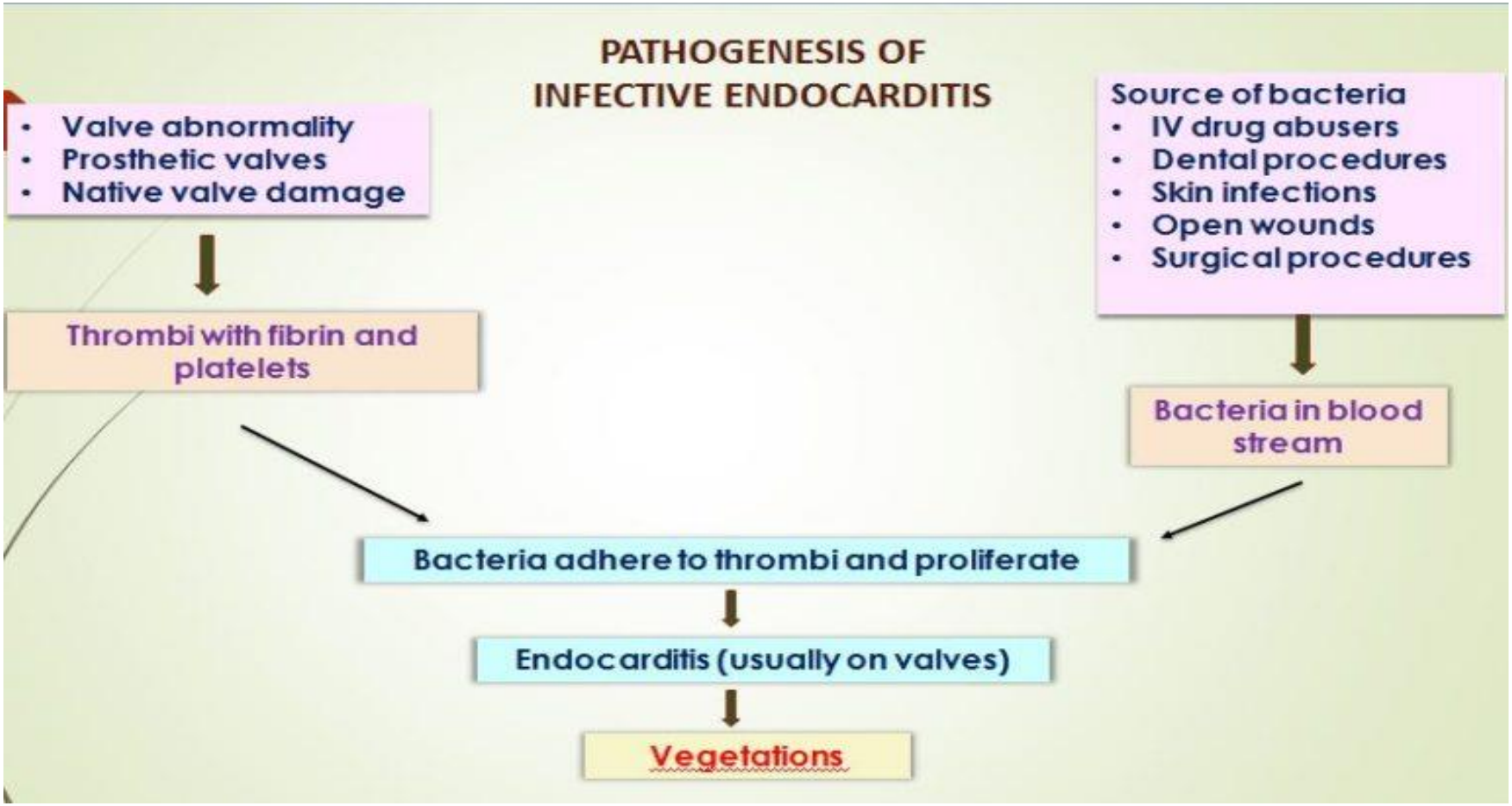
+ previous endocardium affection

1. Rheumatic heart disease (mainly mitral stenosis or aortic stenosis).
2. Congenital heart disease (Small VSD, PDA or bicuspid aortic valve).
3. Degenerative heart disease as calcific senile AS.
4. Prosthetic valve or intracardiac devices.
5. Normal endocardium.


Endothelial Injury
Uninfected Platelet-Fibrin thrombus (NBTE)
Transient bacteremia and attachment at NBTE
Proliferation and pro-coagulant state
Infected, friable, bulky vegetation


a
Pathogens gain (transient) access to the bloodstream
b
Blood flow
Adherence to (injured or inflamed) valve surface
c
Persistence of bacteria
d
Proliferation
e
Dissemination
Activated platelet
Endothelial cell
Bacterium
Subendothelial matrix
Proteins (e.g. fibrin)
Cytokine

Pathogenesis 2



In normal individuals bacteremia disappears soon, but in risk patients, bacteria colonize above the damaged or abnormal valve or defect forming vegetations (thrombi formed of fibrin , platelets and bacteria) and cause inflammation of the endocardium.

Clinical picture 1

1- Constitutional symptoms

- A patient (on risk) with recent history of dental procedure.
- Fever (mild or moderate,< 40)
- Pallor
- Arthralgia & Myalgia.
- Malaise, anorexia and weight loss.

2- Embolization symptoms

a- In cerebral arteries leads to hemiplegia

b- In renal artery leads to kidney infarction

c- In coronaries leads to myocardial infarction.

Clinical picture 2

3- Symptoms of toxemia & immune complex formation

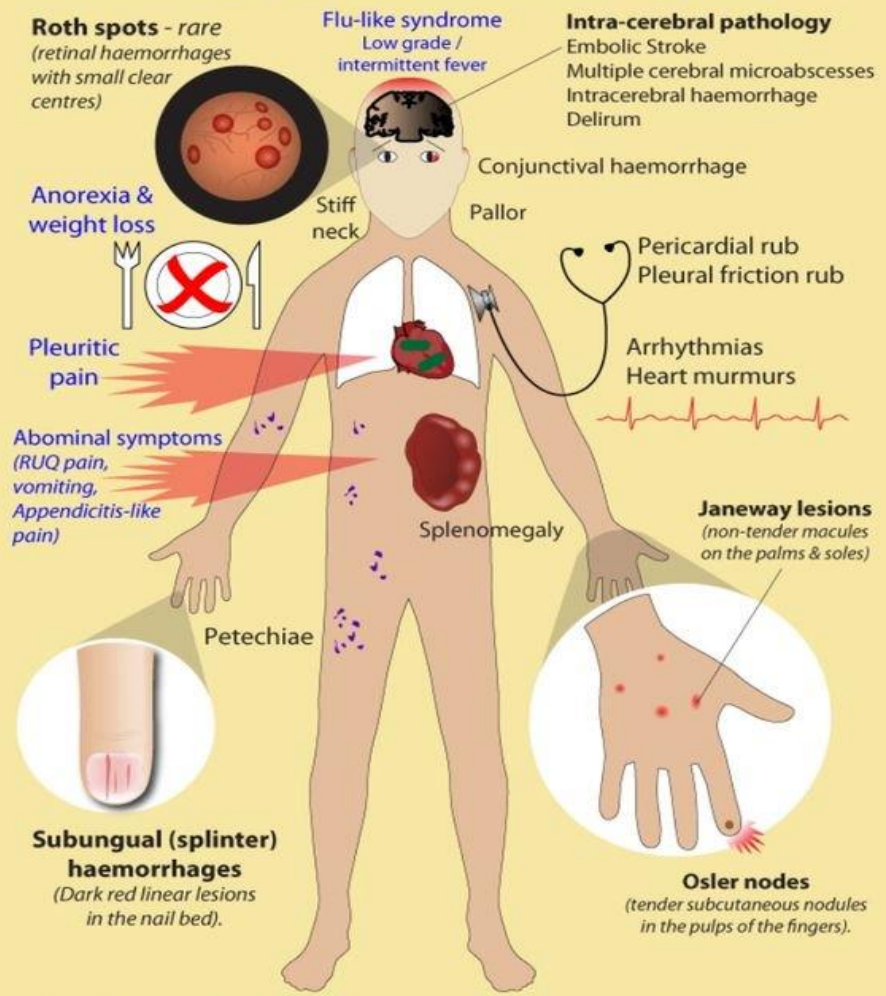
- Mild splenomegaly.
- Clubbing of fingers (pale clubbing) due to toxemia.
- Petechial hemorrhage is common in skin and oral mucous membrane, nails, fundus and conjunctiva.

4- Cardiac manifestations

1. New signs of valve dysfunction.
2. New murmur auscultated by the treating physician.
3. Acute heart failure.
4. Arrhythmia.

Infective Endocarditis

An infection of the endocardial surface of the heart.
Intractable congestive heart failure may result.
If left untreated it is generally fatal.
Subacute endocarditis - symptoms are subtle & non-specific (in blue)



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Investigations

Lab

- CBC: Leukocytosis.
- Elevated ESR.
- Elevated CRP.
- Blood culture is the golden role: better to be taken during peak of fever, 3 samples from different sites within 24 hours and before antibiotic administration.

Imaging

- Echocardiography: direct visualization of the vegetations.

Management (Prophylaxis)1

Who??

- Antibiotic prophylactic therapy is needed in the following risk patients

1. Patients with congenital heart disease (valves or septal defect)
2. Patients with valve disease following rheumatic fever .
3. Patients with prosthetic valve (high risk).
4. History of previous endocarditis.

Dental procedures need prophylaxis ??

- Extractions.
- Periodontal and endodontal (root canal) work.
- Dental implants.
- Initial placement of orthodontic bands.
- Intraligamentary local anesthetic injections.
- Prophylactic teeth cleaning

Management (Prophylaxis)2

Prophylactic regimens for dental, oral, respiratory or esophageal procedures

- Amoxicillin 2g orally in adults and 50mg/kg in children, is recommended 1 hour before the procedure.
- For penicillin allergic patients, azithromycin 500mg; clindamycin 600mg.

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- If the patient is unable to take oral medications (e.g. under general anaesthesia): ampicillin 2g IV or IM is recommended 30 minutes before the procedure onset. For penicillin allergic patients: parentral Clindamycin or azithromycin are used.
- No post procedure dose of antibiotics is needed.
- Oral hygiene is probably more important than antibiotic prophylaxis in preventing endocarditis.

For gastrointestinal or genitourinary procedures

- ampicillin 2 gram + gentamycin 80mg IM or IV are administered half an hour before the procedure.

Management (treatment)

General

- Empirical antibiotic as early as possible.
- Antibiotic choice: bactericidal, Parenteral, high dose and prolonged course (6-8 weeks).
- Later, the patient should be treated by antibiotics according to the blood culture results.
- Delay in diagnosis or ttt = lower survival.
- Surgery: for intracardiac complications.

Antibiotic in use

- Penicillin G + Gentamycin in streptococcal endocarditis.

Others:

- Cefazoline.
- Ceftriaxone.
- Rifampicin.
- Vancomycin (in MRSA).

Thank you